Journal of Surgical Oncology 1997;65:137–140

Thyroid Hemiagenesis

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Background and Objectives: Thyroid hemiagenesis is a rare embryological condition, predominantly in females (3:1) with a left lobe being absent. The associated diseases in the remaining thyroid lobe include benign adenoma, multinodular goiter, hyperthyroidism, chronic thyroiditis, and rarely carcinoma. The most common pathology involved in thyroid hemiagenesis is hyperthyroidism. Presence of carcinoma in a patient with hemiagenesis is quite rare and very few cases are reported in the world literature.

Methods: We report a 30-year-old female who presented with left thyroid mass gradually increasing in size over a period of 3 months. The patient's pre-operative workup included a thyroid scan, which revealed a cold nodule in the left lobe with absent right lobe. A fine-needle aspiration biopsy was suspicious for papillary thyroid carcinoma. The patient underwent thyroid exploration and left thyroid lobectomy.

Results: The operative findings confirmed hemiagenesis of the right lobe and papillary carcinoma in the left lobe. All four parathyroids were in normal position.

Conclusions: The purpose of this presentation is to discuss and review the literature on thyroid hemiagenesis and present a rare case of absent right thyroid lobe with carcinoma in the remaining left thyroid lobe.

J. Surg. Oncol. 1997;65:137–140.

KEY WORDS: **thyroid disease; thyroid cancer; embryologic anomaly**

INTRODUCTION

The common concept of the thyroid gland as a symmetrical structure does not always hold true. Surgeons have recognized that asymmetry is quite common and the right lobe is usually larger than the left. This fact also has been appreciated by physicians experienced in thyroid scanning. Thyroid hemiagenesis, an extreme degree of thyroid asymmetry, is a rare congenital anomaly in which one lobe of the thyroid fails to develop. Isthmus may or may not be present. Embryologically, the thyroid gland develops as a ventral pocket in the midline in the floor of the pharynx. Subsequently, the pocket becomes closed and separated from the pharyngeal wall. The thyroid rudiment is then displaced in a caudal direction until it comes to lay ventral to the trachea by the second trimester of fetal life. Abnormal descent of the gland can contribute to the abnormalities, such as lingual, thyroglossal, cervical or intrathoracic ectopia, and accessory thyroid nodules.

Failure of development of one lobe leading to unilateral agenesis is the rarest of all the anomalies. The cause of unilateral agenesis is not known, but it is believed that the defect may arise from failure of the original anlage to become bilobed and spread out laterally to both sides. Agenesis may be total, unilateral, or isthmic. The pattern of descent gives rise to anomalies that are discussed more frequently, such as lingual thyroid, which is reported to occur in 1 in 3,000 births, but it does not explain the congenital absence of one lobe. In a theory advanced as early as 1949, it is postulated that the unilateral failure of development of the thyroid is related to congenital unilateral absence of thyroid vasculature, but it did not hold true for long, as much contradictory evidence was noted [1]. Vascular anomalies of the thyroid are common in patients with normal bilobar glands (absence of the left

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Accepted 26 March 1997

Fig. 1. Clinically apparent thyroid nodule.

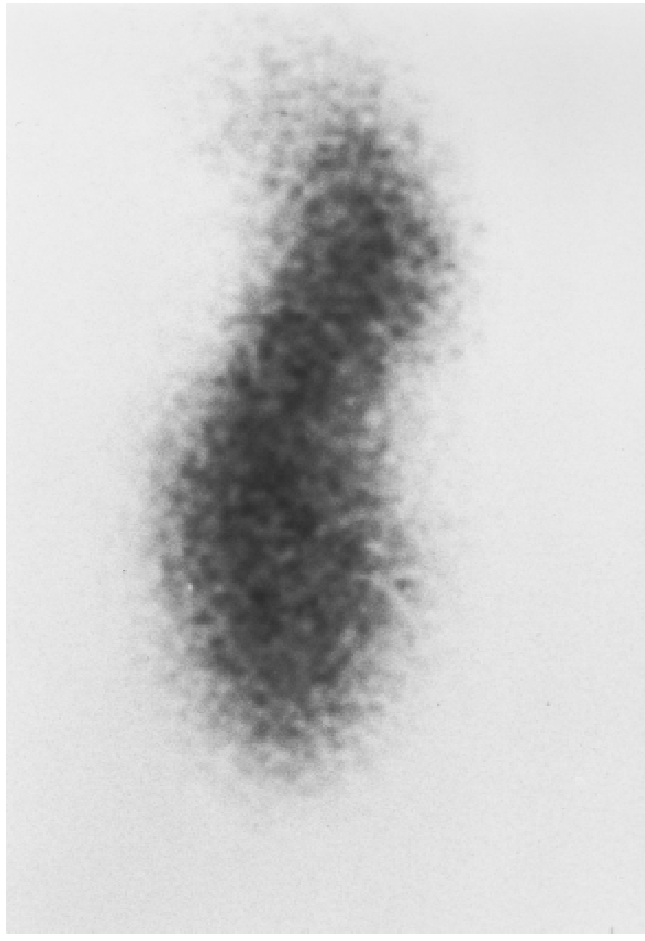

Fig. 2. Thyroid scan showing absent right lobe and cold nodule involving lateral aspect of the left lobe.

inferior thyroid artery in 5 of 100 cases). Some patients with hemiagenesis of the thyroid have had normal vasculature [2]. Congenital absence of other paired organs (e.g., kidneys and lungs) also is more common on left side, again for unknown reasons.

CASE PRESENTATION

A 30-year-old white female was referred for evaluation of a left thyroid mass of 3 months duration (Fig. 1). There was no history of previous radiation to the neck. She did not give any history of other thyroid problems or thyroid dysfunction in the past. Her past medical history was essentially noncontributory, and the patient was placed on Synthroid® 0.1 mg daily. Clinical examination revealed no cervical lymphadenopathy and the trachea to be central. The mass in the left lobe of the thyroid measured ~2 × 2 cm and was quite firm. No abnormality was noted in the right neck and no other masses were noted in the central compartment of the neck. Examination of the oral cavity, larynx, and pharynx was within normal limits with mobile vocal cords. A thyroid scan showed hemiagenesis of the right lobe and cold nodule in the left lobe (Fig. 2). Ultrasound demonstrated a left-sided thyroid lobe with a discrete, solid nodule, and the right neck showed no evidence of thyroid tissue, which was consistent with thyroid hemiagenesis noted on the thyroid scan. The fine-needle aspiration of the left thyroid lobe favored this to be papillary carcinoma. The patient underwent surgery with standard thyroid incision. Intraoperative findings confirmed the presence of a firm nodule in the left lobe of the thyroid and absent right lobe and isthmus. The superior and inferior parathyroids were normally located. A left thyroid lobectomy with removal of all thyroid tissue was performed with the preservation of the parathyroids and their blood supply and safeguarding the recurrent laryngeal nerve on the left side (Fig. 3). Histologically, it was confirmed to be well-differentiated papillary carcinoma. The patient tolerated the surgery well, recovered with no complications, and at present is on replacement therapy with Synthroid® and is doing well.

DISCUSSION

The congenital absence of one lobe of the thyroid is a rare and potentially confusing anomaly seen in fewer

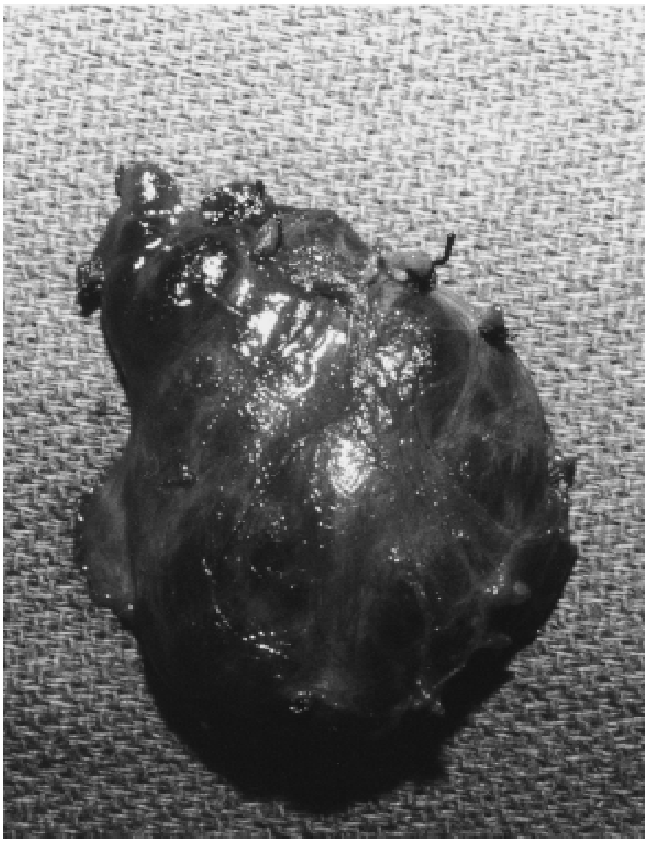

Fig. 3. Specimen after removal of left thyroid lobe.

than 1 in 1,000 patients with thyroid disease. In most reported cases there has been a symptomatic, anatomic, or functional lesion in the remaining single thyroid lobe. Because of the use of thyroid scans to screen asymptomatic individuals with a history of neck irradiation, cases of hemiagenesis have been discovered in which there is no other detectable pathology. A review of the available literature shows that patients with thyroid hemiagenesis are predominantly females (3:1). In addition, the left lobe of the thyroid is absent far more frequently than is the right lobe at a ratio of 4:1. However, the isthmus is absent in an apparently random 50% of cases with thyroid hemiagenesis. Associated diseases in the remaining thyroid lobe include benign adenoma, multinodular goiter, hypothyroidism, chronic thyroiditis, and carcinoma. Some patients were found to be in euthyroid state without any abnormalities. The most common disease of the remaining lobe was hyperthyroidism. The total number of cases of thyroid hemiagenesis is uncertain. The true incidence of thyroid hemiagenesis is difficult to determine since the diagnosis is made in a population being evaluated for some other thyroid pathology. Marshall [3] found one case in 60 autopsies of children. Harada et al. [4] found no cases in 1,007 necropsies. Apart from a few sporadic case reports, the majority of cases have been described by Hamburger and Hamburger [5]. The discovery rate of thyroidal hemiagenesis by imaging has been reported by Maganini and Narendran [6] to be 1 in 1,700 cases and by Hamburger and Hamburger [5] to be 4 in 7,000 thyroid patients. It would appear that the true frequency can be determined only on the basis of large-scale postmortem studies.

The diagnosis of thyroid hemiagenesis depends upon maintenance of an index of suspicion when physical examination or thyroid scan reveals no apparent thyroid tissue on one side. Although hemiagenesis of the thyroid is a benign condition, unawareness of its existence may lead to an incorrect diagnosis and the performance of unnecessary surgery with inherent risks. It is possible to diagnose it clinically when one lobe and the isthmus are absent. Two physical signs may be of help. The edge of the trachea is easily palpable and the edge of the sternomastoid muscle on the affected side is much closer to the midline and overlies the trachea instead of being separated from it. The differential diagnosis would include autonomously functioning nodule with suppression of extranodular tissue, unilateral inflammatory diseases (acute or chronic), metastasis from neoplasm elsewhere in the body, and infiltrative diseases such as amyloidosis or a primary thyroid tumor. A thyroid-stimulating hormone (TSH) test will show no uptake in the case of hemiagenesis and readily demonstrate uptake in the case of suppression of involved lobe in an autonomously functioning nodule. Thyroid scan in a patient with hemiagenesis is quite characteristic and a hockey stick sign may be apparent. The appearance of thyroid carcinoma within the enlarged lobe in this patient is intriguing. Animal data suggest that the high levels of TSH might increase the incidence of thyroid cancer. The increased functional burden caused by the hemiagenetic gland would promote neoplasia. However, the relevance of this is questionable since all the patients with thyroidal hemiagenesis whose TSH levels were measured in Mariani's series [2] had normal TSH value. Even then it is impossible to avoid the suggestion that long-standing elevated levels that can lead to enlargement of the lobe might have played a role in the development of thyroid carcinoma.

Marshall [3] was among the first clinicians to describe numerous anatomic variations of the thyroid, including the clinical entity of hemiagenesis in 1895. Melnick and Stemkowski [7] described the hockey stick sign by imaging study in patients with thyroid hemiagenesis. They also reported four patients and reviewed the world literature on the subject of thyroid hemiagenesis, which revealed a total of 90 cases of thyroid hemiagenesis; however, only 17 of these were reported in the American literature [7]. None of their four patients had thyroid cancer. Sheridan et al. [8] reported a patient with hemiagenesis and Hashimoto's disease. The authors identified and preserved the parathyroid glands in normal position

on the side of the enlarged thyroid lobe; however, they did not identify the parathyroids on the agenic side [8]. The information regarding the parathyroids on the agenic side is not well documented in the literature. In our patient, we were able to identify all four parathyroids, which were in normal position, even on the agenic side. Embryologically, the parathyroids develop from the pharyngeal pouches, the superior glands arising from the 4th pharyngeal pouch and the inferior glands arising from the 3rd pharyngeal pouch. The inferior parathyroids descend with the thymus and occasionally lie in the thymic capsule. Most often they separate from the thymus and lie on the posterior surface of the inferior pole of the thyroid along the inferior thyroid artery. Piera et al. [9] reported three cases of thyroid hemiagenesis in 1986; however, they documented the normal presence of parathyroids on the side of the enlarged thyroid lobe only in one case. It is important for the thyroid surgeon undertaking surgery on a hemiagenic thyroid to appreciate the position of the parathyroids and to make every effort to preserve the parathyroids on the side of the thyroid lobectomy.

McHenry et al. [10] recently reported seven patients with thyroid hemiagenesis—a collected experience of five physicians. They reported four female and three male patients ranging in age from 17 to 58 years. The pathologies included follicular adenoma, Graves' disease, and nodular goiter. One patient had follicular carcinoma of the thyroid. They emphasized the need for preoperative recognition of thyroid hemiagenesis in order to make critical decisions regarding surgical intervention. McHenry et al. [10] suggested that all patients with thyroid hemiagenesis who do not have indications for surgery should have monitoring of their thyrotropin levels, treatment of thyrotropin elevation with thyroid hormone, and careful followup evaluation of the development of neoplastic disease. Our patient is a unique case of an absent right thyroid lobe in a 30-year-old female with papillary carcinoma in the remaining left thyroid lobe and all four parathyroids present in normal position.

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